CLINICAL VIGNETTE

An Unusual Bruise: A Case of Achenbach Syndrome

Jarod DuVall, MD and Bryan Lopez, MD

Case Presentation

A 37-year-old woman without any significant past medical history presented to ambulatory clinic for evaluation of a spontaneous bruise on the volar aspect of her right 4th digit. This occurred the night prior to her office visit while she was typing on her computer. She initially noted pain and warmth in her finger and subsequently developed the bruise.

She reported two similar episodes in the past. The first occurred 4 years prior and involved bruising in multiple fingers. The second episode was identical to the current presentation. Both episodes involved mild pain followed by bruising minutes to hours later. She denied any past blood clots or bleeding problems including abdominal bleeding after injuries, GI bleeding, hematuria or menorrhagia.

She reported a history of suspected Raynaud phenomenon. Her mother had primary Raynaud phenomenon, and the patient reported that her fingers turned white when exposed to cold. They did not turn blue, and she had no prior finger ulcerations or gangrene.

She did have intermittent pains in her hand joints but believed these were related to her work as a ceramic restorer and were not out of proportion to her peers. Otherwise, she was free of significant joint pains or joint stiffness. She reported a sun sensitivity rash and mild dry eyes. She had no connective tissue symptoms including skin thickening, dry mouth, oral ulcers, hair or nail changes, or other rashes.

At work, she sometimes used a vibrating tool, but she exclusively used the tool in her left hand while the bruising always occurred in her right.

Her only medication was a combined oral contraceptive pill. She denied tobacco use but did occasionally use alcohol and marijuana.

Her vital signs were unremarkable. Her physical exam was notable for a normal cardiopulmonary exam, normal radial pulses, normal bilateral grip strength, and intact sensation to light touch in her fingers and hands. An ecchymosis centered around her 4th distal interphalangeal joint on both dorsal and palmar aspects was present. There was no significant synovitis of any hand joints or wrists.

Labs included normal complete blood count, comprehensive metabolic panel, PT/INR/PTT, TSH, factor VIII activity, von Willebrand Factor activity, hepatitis C, and HIV. Her ANA was positive at 1:80 in a homogenous pattern. Otherwise, she had a negative rheumatologic work-up including RF, CCP, dsDNA, complement levels, c-ANCA, p-ANCA, myeloperoxidase antibody, proteinase-3 antibody, SSA, SSB, centromere B antibody, SM antibody, RNP antibody, and Jo 1 antibody. X-ray of her right hand was unremarkable.

Discussion

Initially, there was concern for an underlying vasculitis given the patient's spontaneous hematoma with history of Raynaud's phenomenon. She also mentioned possible easy bruising though no other significant bleeding history. Additional testing was unremarkable. Though Raynaud disease can be associated with an acute blue finger and less commonly with significant ischemia, the patient's episodes had no relation to her Raynaud's symptoms. Her presentation was most suggestive of Achenbach Syndrome, which the patient herself diagnosed during follow up correspondence.

Achenbach Syndrome, which has been called Spontaneous Blue Finger Syndrome, is a clinical phenomenon first described in 1958. It involves bruising of the fingers, usually the volar aspect, without a known etiology.[1] This can occur spontaneously or with mild trauma and can be associated with sensations including pain, swelling, numbness, and coldness occurring minutes to hours before bruising develops.[1] There are no known complications, and the disease is considered benign.[1]

Though no cause is known, there is thought to be an underlying vascular etiology. One study demonstrated presence of capillary microhemorrhages[2] while another found skin biopsies with changes suggesting skin and microartery repair after a hypoxic or ischemic insult.[1] No findings were suggestive of a thromboembolic, atheroembolic, or vasculitic process.[1]

While there have been no large studies of Achenbach Syndrome, there have been a number of case reports and a case series. A 2012 report of a woman with multiple episodes of nontraumatic finger discoloration with tip sparing.[3] Her evaluation was remarkable for a mildly positive ANA (1:40) and negative RF, CCP, and anti-SCL 70. She also had a normal TTE and a normal CT angiograph of the aortic arch and right

upper extremity. This patient received aspirin and later dipyridamole until the diagnosis was made.

While Achenbach Syndrome is a benign condition, it is important to rule out other causes of blue fingers especially acute digit ischemia. The differential for blue digits includes Raynaud phenomenon, thromboangiitis obliterans, cryoglobulinemia, collagen vascular diseases, polycythemia, venous thrombosis, vasospasm, vibration injury, atherosclerosis, emboli, trauma, thoracic outlet syndrome, and frost bite.[4]

A case series at a tertiary hospital's vascular department identified 22 patients with acute blue fingers and normal pulses and symmetric upper extremity blood pressures. None of the patients had prior episodes. Only five patients were found to have an underlying diagnosis that could explain symptoms including two patients with Raynaud's disease, two with polycythemia, and one with thoracic outlet syndrome. All but three patients underwent additional studies, which were unremarkable. Imaging included arterial duplex evaluations of the upper extremity, transthoracic echocardiograms and 24 hour heart monitoring. Twenty one patients were re-evaluated at 6 weeks and only 5 had symptoms. At longer term follow up, only 3 of 19 patients had recurrent symptoms, two of which had polycythemia. One of the polycythemia patients had a subclavian thrombosis. No patients had embolic events on re-evaluation. Many of the patients were maintained on anti-platelet therapy. The authors concluded that an acute blue finger appears to be a benign condition and additional work-up beyond hypercoagulable or collagen vascular diseases is unnecessary.[4]

Of note, a letter to the editor in response to the above publication advised caution and discouraged physicians from considering acute blue fingers to be a benign condition. The authors reported a patient later found to later have left subclavian artery stenosis requiring distal phalangeal amputation. However, that case involved all 5 fingers including finger tips.[5]

Conclusion

Achenbach syndrome is a benign condition not commonly encountered. Physicians should be aware of the condition to provide reassurance and avoid unnecessary testing and treatment such as antiplatelet medications. The disease tends to spare finger tips and does not include splinter hemorrhages—which would be more suggestive of an acute ischemic or embolic process and would warrant additional evaluation. A thorough history and physical examination are important to uncover any symptoms or signs suggestive of an underlying vasculitis or an acute ischemic event.

Figures

REFERENCES

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